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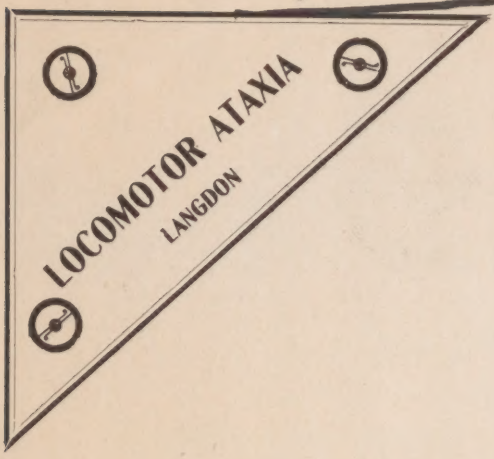

LOCOMOTOR ATAXIA

AS IT ESPECIALLY INTERESTS THE GENERAL PRACTITIONER *

BY

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As the most common chronic disease of the nervous system, Locomotor Ataxia may be said to interest the practitioner of medicine

First: as regards its causation and pathology.
Secondly: as regards its diagnosis and prognosis.
Thirdly: as regards its treatment.

Its literature is voluminous; comprising, up to 1900, some 2000 essays (Mobius).[3]

A paper intended for a gathering such as this, of practitioners who meet to compare notes, correct errors, and perhaps imbibe a few new ideas from each other, should be, if my conception is correct, a concise, up-to-date presentation of the salient points of the subject, rather than a formal and more complete didactic discourse.

When the commander of a modern battle-ship prepares to engage the enemy, one of the first orders given is to "strip for action," in other words to clear the decks of all superfluous and for the time being, useless material; which may be not only useless, but actually dangerous or detrimental to success in the coming battle.

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CURRENT FALLACIES CONCERNING THE DISEASE.

Applying this principle to the subject before us, suppose we clear the clinical and pathologic fields of various fallacies respecting the disease known as locomotor ataxia since its classic description by Duchenne in 1858.

One fallacy that has been pretty generally cast aside, is the idea that Locomotor Ataxia is primarily or chiefly, a disease of the Spinal Cord. It is hardly necessary in these post-neuronic days, to direct attention to the fact that the neurons usually and mainly affected in this disease, are situated, so far as their main masses, namely their bodies and nuclei, are concerned, in the ganglia on the posterior roots of the various spinal nerves, and thus are entirely outside the cord. Incidentally, it is true, these neurons send processes (fibres) into the cord, where they transverse chiefly the columns of Goll and Burdach. They also however send other processes equally important, to the periphery of the body, which processes are necessary to the conduction of sensations of touch, pain, temperature, posture, etc., from the skin, muscles and joints.

The *peripheral sensory neuron* therefore, as one of these entire anatomical units is called, forms a continuous conducting medium for various sensory impressions, beginning at the periphery and ending at the oblongata; with various side branches (collaterals) passing off at different levels of the cord, to influence the motor neurons of the anterior horns, and so develop the phenomena of "reflex" action. It so happens therefore, when this set of neurons, in part or as a whole, "degenerates," as we technically term the process of molecular death with failure of repair, the examination of the Spinal Cord reveals a "lesion," (a "sclerosis" to use another fallacious term), in the situation once occupied by the processes of these now defunct neurons. That is to say, a lesion in the columns of Goll and Burdach with more or less disseminated lesions in other situations once occupied by collaterals and connections of the same neuron groups.

But, so do some of the peripheral nerves reveal a lesion, when examined, which they seldom are. Hence, why not call the disease a "peripheral nerve disease," which should be quite as correct as to say a "spinal cord disease."

This concentration of attention on spinal cord changes, and the ignoring of equally important alterations in the peripheral nerves, has been one of the curiosities of pathology and morbid anatomy of the past. And even today, many of our

current text-books describe the pathology of the disease as a "sclerosis of the posterior columns of the cord."

It is as if we were to call a cerebral hemorrhage, in the motor region, a "spinal sclerosis," because the autopsy finally reveals a degeneration in the pyramidal tracts of the cord. We may accept as established therefore, the fact that Locomotor Ataxia is, pathologically speaking, a degeneration in various peripheral sensory neurons, belonging to the posterior root ganglia chiefly and that sclerosis of the columns of Goll is merely an incident indicating the death of the axons of these neurons and their replacement by a new growth of neuroglia tissue.

The third fallacy of which we need to divest ourselves, is, that the presence of Locomotor Ataxia implies the certainty of a previous syphilis,acquired or hereditary.

This is undoubtedly an error, which, aside from doing injustice to some patients and injury to others, has probably been directly responsible for more mistakes in therapy, than any other one idea of the disease.

It is only necessary to refer to the fact, that so conservative an observer as Sir William Gowers has stated, that, "In a few cases, less than ten per cent. of the whole, it (syphilis) can be excluded with confidence." Other writers concede a considerably larger percentage as non-syphilitic, even up to forty per cent.

The writer has distinctly in mind at the present writing, two well-marked, typical cases of tabes, one in a physician, whom he has known from youth, and in whom syphilis can be satisfactorily excluded. The other, a motherly old lady with healthy children, who has atrocious lightning pains, loss of pupillary light reflex and of knee-jerks, with ataxia of gait and station, who presents neither symptoms nor history of syphilis.

These cases might be multiplied, although we all know how difficult it is to exclude the possibility of accidental syphilis.

A fourth fallacy, even more important to recognize, and dependent on the preceding, is the idea that anti-syphilitic treatment is useful in the cases which *have* a history or evidence of previous syphilis.

The concensus of neurologic opinion, at the present day, is that such treatment is not only useless, but is actually in many cases harmful.

The reason for this is evidently the fact that tabes is not an exudative inflammatory process, but a degeneration, and it is only in the former process (i.e., inflammatory exudation) that mercurials and iodides can be expected to exert any good effects.

Having cleared the clinical and pathologic fields of the foregoing fallacies, we may now take up the question of

DIAGNOSIS.

How early and by what symptoms may we positively diagnose Locomotor Ataxia? is a question often asked.

Since the essential pathologic element of the disease is a premature molecular death of sensory neurons, it is natural that we should look to the sensory functions for the first symptoms.

It may help us here to remember the pathologic law, that the longer the duration of a function in the biologic series, the more persistent, or resistent, are the structures which manifest it, under adverse influences.

Or, per-contra, the later the development of a function, biologically speaking, the more vulnerable it is to disease. We have only to bear in mind here, the readiness with which aphasia and other disorders of speech (a late acquirement) are produced, as compared with the persistence of ordinary motion. Also the ease with which the central inhibitory functions in many persons, disappear under the toxic influence of small doses of alcohol and other poisons.

Applying this principle to the peripheral neurons subserving sensory functions, it is evident at a glance, that contact or tactile sensation, is the most elementary, and consequently most ancient form of sensory cognition.

Sensations of pain, of heat and cold, of muscle sense, posture, articular sense, all appear later, and in their full perfection, are the result of educational acquirements. Hence, when the old man begins to fail in his neurons, he becomes less precise in automatic movements, which depend on nicety of articular and muscle sense. He is also commonly less sensitive to degrees of heat and cold, and even to pain. The same order of events obtains when the neurons die slowly from other causes than age; toxins for example, or arteriosclerosis diminishing the blood supply.

Consequently we find as the earliest signs of tabes, delay in transmission, or actual loss of pain, temperature and muscle senses, and these signs are naturally manifested in those neurons, whose processes are longest, and whose endings consequently, are farthest removed from the source of nutrition, the neuron body. In other words those which end peripherally about the feet and hands.

The question therefore, "how early may we make a diagnosis of tabes?" may be answered; "Probably long before the patient complains of any symptoms;" provided we were to examine him carefully, which of course we seldom do-at such a time. Practically however, the first symptoms, which lead, or should lead, to a careful examination and a correct diagnosis, are the "lightning pains," due to beginning degeneration in pain conducting fibres; and at such time, not only will the pains be present, usually above and below the knee, but if we examine the ankles and feet, even where pains are absent, in these regions, we will find almost invariably, *absolute loss of pain sensibility*, (analgesia), occuring in patches, indicating areas where certain fibre endings have been absolutely destroyed.

This is a very simple test to make in doubtful cases. We simply test the patient, with his eyes closed, about the ankles with the head and point of a pin, alternately, and ask him to say "sharp" or "dull" at each touch; While the "touch" may be felt every time, the "quality" of the sensation will not, and he will say "dull" frequently when the point penetrates the skin; the same symptom is often present about the wrists.

At the time of this "patchy" loss of pain sense, there will also be found, usually, more or less thermo-anesthesia as well. Naturally if the knee-jerks and the pupillary light reflex be lost, there is no longer any doubt about diagnosis, but it is in the very cases where the knee-jerks are still present and the pupils respond more or less readily to light, that the sensory symptoms above noted, about the ankles and wrists, are of great value.

Another sensory symptom, very frequently present, is a *thoracic band* (sometimes abdominal) of *tactile* anaesthesia. A peculiarity of this band is that it is never symmetrical on the two sides, but is a little higher or lower, or a little narrower, on one side.

The width of this band may vary from half an inch to six inches. Its asymmetery is due, probably, to the fact that it represents the distribution of one or more posterior root ganglia, which are unequally developed, or unequally degenerated, or both, on the two sides of the body.

While therefore, the four cardinal symptoms; lightning pains, lost knee-jerks, lost light reflex, and ataxia of gait, are diagnostic beyond a doubt, a strong presumptive diagnosis may be made by the cutaneous sensory symptoms alone, provided a multiple neuritis and a focal cord lesion be excluded. As both of these conditions have a definite period of onset, while tabes has not, the distinction is comparatively simple.

It is not sufficient however, to make a diagnosis of Locomotor Ataxia. The symptoms may be unmistakeable, the diagnosis undoubted, and the patient still have another disease *in addition to Tabes*. I refer of course to *Paresis*.

While it is reasonable to assume that this association means the involvement of cortical neurons in the same degenerative process, yet this is not yet proven. The vastly greater importance however of any involvement of the psychic functions, is such as to demand a most careful examination of the mental status in every case of Locomotor Ataxia.

PROGNOSIS.

Having arrived at a diagnosis, the question of prognosis naturally arises.

The writer in a previous paper [2] has said: "The prognosis of locomotor ataxia is not by any means so hopeless as is generally taught. Most cases not bed-ridden are susceptible of distinct improvement."

This view is sustained by distinguished authorities:—Mobius [3] for example has said: "In so far as the peripheral nerve fibres are primarily affected in tabes, the tabetic symptoms are curable, * * * *" and again, "In any case the physician does well not to express himself too pessimistically. The patient has a right to encouragement and the physician may point out the fact that the disease may improve or remain stationary."

Collins[4] likewise states: "It should not be forgotten that, clinically, tabes is a recoverable disease. Unfortunately recovery does not often occur." In general terms, we may say *the earlier the diagnosis, the more favorable the prognosis*. Hence a very practical division of cases, as regards prognosis, would be, into three groups.

First: Those diagnosed very early with comparatively small extent of damage to sensory neurons; cases with recently developed lightning pains, numbness or other paraesthesiae of feet and legs; analgesia about the ankles, perhaps diminished and unequal knee-jerks; little or no ataxia; with sluggish pupils but not complete loss of light reflex. In these cases other things being equal, arrest of the disease or a very material slowing of its progress may be reasonably expected. I have had such cases under observation for many years, with no

(2) New York Medical Record, January 8, 1898.
(3) Mobius, Paul J.: Tabes dorsalis; Twentieth Century Practice of Medicine, Vol. XI.
(4) Collins, Joseph; Treatment of Nervous Disease, Wm. Wood & Co., New York, 1900.

perceptible progress, and even with apparent improvement, so far as general health and ability to make a living are concerned.

A second group comprises the fully developed cases where symptoms have existed for five or ten years and even longer; cases with lightning pains, ataxia of gait and station, lost knee-jerks and iridoplegia to light. A noticeable fact concerning the pains, is, that they commonly accompany exposure to cold or sudden lowering of temperature or barometer. They also commonly attend any gastro-intestinal "upset," as a so-called "bilious attack" etc. In fact I have some patients whose standard remedy for the pains, and a very effective one, is a dose of Castor Oil; others are greatly benefited by a purgative dose of Sodium Phosphate.

Toxaemia of intestinal origin therefore, would seem to be an important factor in excitation of the pains.

In these fully developed cases, the prognosis is naturally less favorable, since the patient has lost large groups of sensory neurons. If the degeneration has been rapid and hygiene defective, inflammatory complications (neuritis) are apt to arise. The most that can be hoped for in these cases, therefore, is a degree of comfort, depending on the extent to which the patient can adapt his life to the lessened capacity of his sensory nervous system. So long as the patient can get about however, and has means and disposition to take proper care of himself, his existence may be made quite comfortable, even in these fully developed cases.

In the third group of cases are the helpless, the bed-ridden, with complicating lesions on the part of the motor apparatus, (anterior horn degenerations, etc.) Those with arthopathies, bladder defects, laryngeal, gastric, vesical and rectal "crises" etc. Symptomatic and palliative measures, are of course the only resource here.

So far, any reference to optic nerve atrophy, with its distressing accompaniment, gradually progressive loss of vision, has been purposely omitted.

About ten percent of tabetics, present optic atrophy, and it is a noticeable clinical fact that these are seldom the worst cases, as regards bodily comfort and nutrition generally. In fact, the statement is dogmatically made in some text books, that "optic atrophy arrests the ataxia." This is not only a self-evident fallacy, but one of such absurdity that it is remarkable it should have gone unchallenged for so many years. The only rational explanation of the slow progress, and even improvement, in many cases after the development of blindness, is the

tact that the patient is necessarily less active, *the element of enforced rest is the important factor.* This should and does furnish us a valuable hint in the clinical management of all cases.

TREATMENT.

My remarks on treatment will be necessarily brief. Widely distributed groups of sensory neurons are undergoing the slow molecular death, which we call degeneration. The indications are plain—to promote by every means in our power, the nutrition of these failing structures. No medicines are equal to rest and feeding.

The tabetic patient is one-half, one-third, one-fourth, or less, of a man, according to the amount of destruction in his sensory neurons and the exigencies of his occupation.

To diminish function, is to lessen waste; hence, rest, a mild climate, an even temperature, avoidance of dissipation, care in diet and hygiene, are all obviously indicated. Certain drugs have a food value. Iron, the phosphatic compounds, as Lecithin, Glycero-phosphates, etc. Strychnine has little value unless to meet some indications on the part of the heart, or perhaps the bladder. Mercury and iodides, as before intimated, are useless and often harmful.

Symptomatic treatment is often necessary. For the pains, in addition to the mild laxatives, already mentioned, there is no better routine prescription, in the writer's experience, than *Aluminium Chloride*, in doses of three or four grains dissolved in half a tumblerful of water, three times daily, as first recommended by Gowers. Atropine, hypodermatically, is of value in the various "crises."

Popular remedies containing coal-tar products, are much used by the laity. They are probably all harmful if long continued; and sometimes contain opiates, also objectionable.

In advanced cases with crises and severe pains, comfort is to be secured at any cost, but the resort to narcotics should be delayed until absolutely necessary. For the ataxia, the "compensation" method of Fraenkel, or treatment by co-ordination exercises has a definite value in selected cases. Its chief drawback, and a very serious one, is the difficulty of inducing the average patient to persist in its use, unless he be under the constant care of the physician or a skilled attendant. The details of its use may be found in most modern text books on nervous diseases.

Electricity appears to be of benefit in a considerable proportion of cases.

The writers experience has led him to value highly that current known as "sinusoidal"—a current of high voltage, small amperage and rapid alternation (480 to 1900 alternations per second). Its application is simple and very agreeable to the patient. A foot plate and broad spinal electrode are used to administer it, the application being made every alternate day for about six weeks; then omitted for a month or two, and resumed for another six weeks.

Under its use and appropriate hygienic, dietetic and supporting treatment, the lightning pains commonly cease, the ataxia improves, and the patient gains in general nutrition.